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Polyostotic Coexisting Fibrous Dysplasia and Aneurysmal Bone Cyst of the Chest Wall

F. Tresserra, R. Rami[1], A. Salas, A. Domingo, P. Forcada, G. Gonzalez-Pont, X. Tarroch, and M. Cuesta[1]
Departments of Pathology and [1]Thoracic Surgery of Hospital Mutua de Terrassa, Terrassa (Barcelona), Spain

Summary

A case of a 39-year-old woman with a palpable mass in the right hemithorax is presented. The mass had been growing during the last 16 years. Radiographs and computed tomography showed two lesions in the right thoracic wall: the greater was in the anterior and lateral portion of the 7th rib, the minor lesion in the 6th rib costovertebral joint. Both lesions were surgically removed. Histological examination demonstrated the association of fibrous dysplasia and aneurysmal bone cyst in the two lesions. The coexistence of these two lesions supports the theory that aneurysmal bone cyst may repreent a secondary change due to haemodynamic alterations of the vascular bed caused by fibrous dysplasia.

Key words

Aneurysmal bone cyst – Fibrous dysplasia – Chest wall tumors

Dysplasia fibrosa kombiniert mit einer aneurysmatischen Knochenzyste

Es wird über eine 39 Jahre alte Frau mit einer tastbaren Tumormasse auf der rechten Thoraxseite berichtet. Die Masse ist im Laufe der vergangenen 16 Jahre allmählich größer geworden. Die Röntgenuntersuchung und der Thorax-Scanner hatten zwei pathologische Befunde in der rechten Thoraxhälfte entdeckt: der größere auf dem vorderen und seitlichen Teil der 7. Rippe, der kleinere im Bereich des costovertebralen Gelenkes der 6. Rippe. Beide Geschwülste wurden chirurgisch reseziert. Bei der histopathologischen Untersuchung stellte sich heraus, daß es sich um eine Dysplasia fibrosa und eine aneurysmatische Knochenzyste handelte. Die Koinzidenz dieser beiden Befunde bekräftigte die Theorie, daß die aneurysmatische Knochenzyste einen sekundären Zustand der Dysplasia fibrosa durch haemodynamische Veränderungen der Gefäße darstellen kann.

Introduction

Aneurysmal bone cyst (ABC) was first described by *Jaffe* and *Lichtenstein* in 1942 as a non-neoplastic bone lesion (11). A few years later, *Jaffe* pointed out the possibility that ABC might sometimes represent a secondary growing in a preexisting bone lesion. Since then many reports have appeared in the literature associating ABC with other bone affection: non-ossifying fibroma, chondroblastoma, giant-cell tumor, osteoblastoma, fibromixoma, hemangioendothelioma, fibrous histiocytoma, fibrosarcoma, osteosarcoma, and fibrous dysplasia (FD) (13, 19). Fibrous dysplasia was described by *Lichtenstein* in 1938 as a benign bone mesenchymal maldevelopment (12). The association of FD and ABC is rare but has been previously documented (1, 3, 4, 6, 8, 9, 13, 14, 16, 17, 18). Here a case of polyostotic form of coincidental fibrous dysplasia with aneurysmal bone cyst is described. To our knowledge this is the first report of the association of this two lesions in two different places.

Case report

This patient is a 39 year-old female with a history of epilepsy. Sixteen years before the present admission, the patient underwent an biopsy of a right rib seen on a chest radiograph to be widening. The specimen obtained was normal bone tissue and no further procedures were performed. The patient is a non-smoker and takes no medication, except for anti-epileptic drugs.

The patient reported that, along the years, she had been feeling a non-tender, hard, progressively growing mass under the scar left by the rib biopsy. On physical examination, a 10×10 cm hard, non-movable mass could be seen and palpated on the antero-lateral aspect of the right chest wall, under the breast. The mass was firmly adhered to the chest wall. The overlying skin did not show any abnormality and did not adhere to the mass. Slightly above the mass, the scar from the previous operation looked completely normal. Results of the rest of the physical examination were normal.

Complete blood count, coagulation, and blood-chemistry analyses were within normal limits except for the alkaline phosphatase, which was 1418 IU/L. EKG and pulmonary function tests were normal.

Postero-lateral and lateral chest radiographs showed a multilobulated, well-defined mass on the right hemithorax, arising from the 7th rib, which was partially substituted by the mass. This mass had two different components: one extrathoracic, of lesser volume and very calci-

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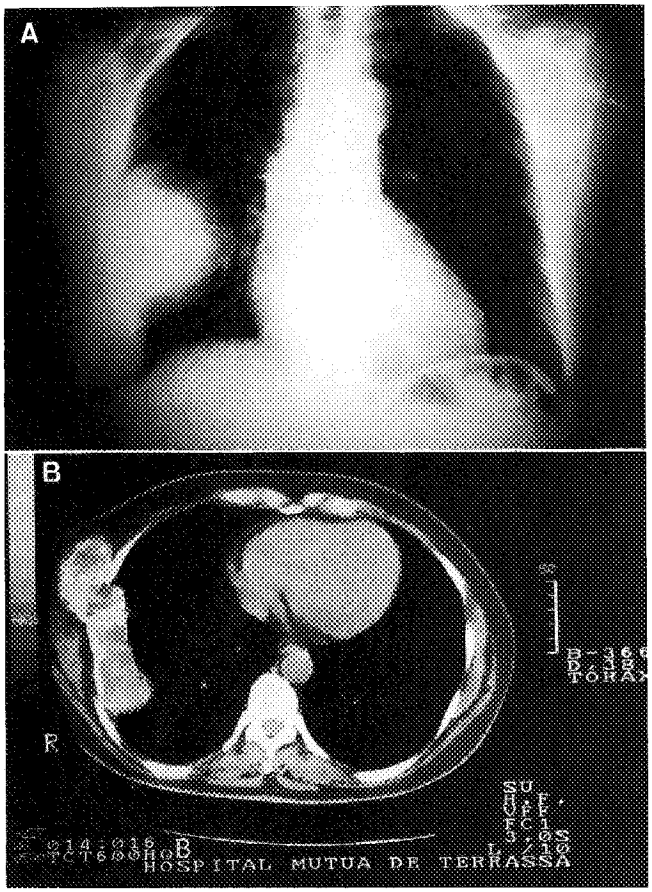

Fig. 1 a Postero-anterior chest wall radiograph showing multilobulated well defined mass in the right side.

Fig. 1 b Computed tomography showing the extra- and intrathoracic components of the greater mass. Note the peripheral calcifications.

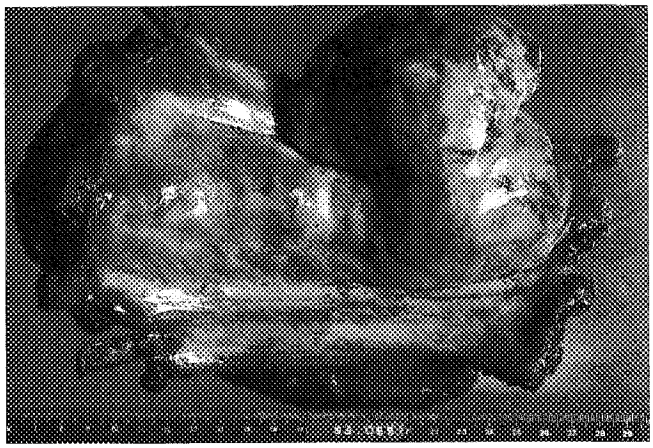

Fig. 2 Gross appearance of the mass at the 7th rib in its intrathoracic site.

fied; and one endothoracic with less calcifications. Computed tomography scans of the chest showed an expansive lesion on two different locations: at the costovertebral join of the 6th rib and along the lateral and anterior portions of the 7th rib. The lesions contained peripheral calcifications, mainly on the extrathoracic component and septa, were well-defined, and did not affect the lung parenchyma (Fig. 1). Bone scan revealed a bilobulated, homogeneous, intense uptake on the 7th rib. This examination did not show other skeletal alterations.

The patient underwent a right postero-anterior thoracotomy. The latissimus dorsi muscle was detached from the spinal and pelvic insertions and spared for chest wall reconstruction. The serratus anterior muscle, which adhered to the chest-wall mass, was completely freed from its insertions and left attached to the mass for en bloc resection. The anterior segment of the 5th rib and the anterior and lateral segment of the 6th, 7th, 8th, 9th and 10th ribs were removed en bloc with the tumor and serratus anterior muscle, and a wedge resection was carried out of the middle lobe, which was firmly attached to the intrathoracic component of the tumor. Then the posterior segments of the 5th and 6th ribs were excised, including the costo-vertebral component of the lesion. The chest-wall defect was repaired with a GORE-TEX mesh, sutured along the periphery of the defect. The anterior and lateral portions of the mesh were subsequently covered with a pedicled latissimus dorsi muscle flap; and the posterior aspect was covered with the inferior segment of the trapezius muscle. Two chest tubes were left in the pleural space; and five low-suction tubes were inserted between the mesh and the muscular cover, and between the skin and the muscular layer. Subcutaneous tissue and skin were closed by direct suture using absorbable material. The postoperative course was uneventful.

Grossly the surgical specimen was composed of a part of the chest wall including five ribs. The intrathoracic side showed a bilobulated yellowish tumor growing along the 7th rib trajectory measuring 13×10 cm. Beneath the muscle covering the extrathoracic side, there was a nodular formation of 6 cm in greatest diameter (Fig. 2). The cut surface of the tumor presented a rather firm, whitish or pink-grey tissue with haemorrhagic areas involving and destroying the 7th rib. The other ribs were not affected by the tumor.

The lesion in the posterior portion of the 5th and 6th ribs measured 2×1.5 cm and showed similar characteristics.

Portions of both specimens were fixed in 10% formalin, decalcified in 5% formic acid and embedded in paraffin, sectioned in 5 µm slides, and stained with hematoxilin-eosin.

Microscopically (Fig. 3) both lesions arose from the ribs, breaking the cortex. They were composed of variable-density fibrous tissue, involving narrow, curved, sometimes with "c" or "s" shapes, and misshaped bone trabeculae in different ossifying states, all findings characteristic of fibrous dysplasia. In abrupt transition with these zones there were areas with mesenchymal tissue and anastomosing fibrous-walled channels containing blood. These channels did not contain elastic lamina or muscle layer but were lined by endothelial cells. The fibrous wall contained red blood cells, hemosiderin deposits, and spicules of reactive bone. A rich benign-appearing giant multinucleated cell infiltration was seen. These features were consistent with aneurysmal bone cyst changes. No atypical mitotic figures were observed. The diagnosis of FD with ABC in both lesions was made.

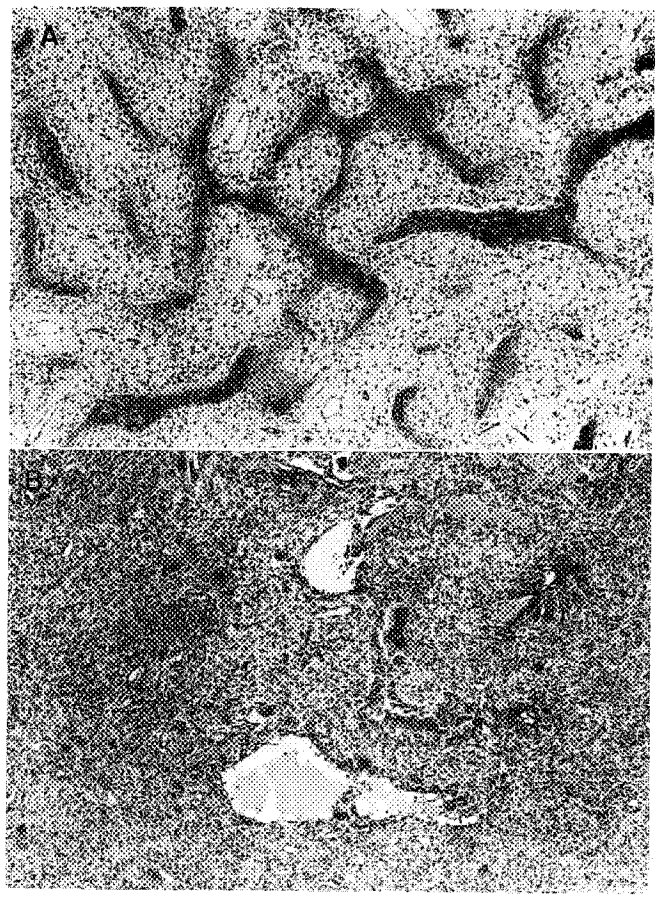

Fig. 3a Microscopic image of a zone with curved and misshaped bone trabecules surrounded by stromal tissue characteristics of fibrous dysplasia (H&E × 100).

Fig. 3b Aneurysmal bone cyst is microscopically characterized by vascular spaces cystically dilated. Many giant multinucleated cells were seen in the stromal tissue around the vessels (H&E × 100).

Discussion

The majority of the tumors of the chest wall are malignant or metastases (15): fibrous dysplasia is with the enchondroma one of the most frequent benign tumoral lesions of the chest wall (5). It may appear as a solitary focus or may be multiple. A monostotic variety occurs in older children and young adults, affecting ribs, femur, tibia, jaw, and calvarium (10). The less common polyostotic type has usually unilateral distribution of the lesions, and frequently in association with endocrine dysfunction, precocious puberty in the female, and cutaneous hyperpigmentation (Albright's syndrome) (4). Malignant change occurs in 4 to 10 % of cases (7). FD has been associated with osteosarcoma, chondrosarcoma, and spindle-cell sarcoma (10, 20). The risk of malignant transformation increased with radiation therapy (3). ABC is probably a secondary "blowout" bone lesion, associated with many bone tumors or disorders (13). Initial symptoms include pain, rapid enlargement and possible skeletal or neurological dysfunction, depending on the location and extent of the bone destruction (3). The coexistence of FD and ABC has been reported in multiple locations (Table 1), but always as a monostotic lesion. The case we describe had both lesions coexisting in two different places. This fact supports the possibility that the origin of ABC is in some anomaly of the vascular bed of a given area, as some authors have pointed out (4, 8): mechanical closure of the vessels in the fibrous tissue of FD could alter the hemodynamism of blood supply, creating abnormal vascular channels that appear to be an arteriovenous fistula. The fistula would erode the adjacent tissue, producing reactive giant cells and active stromal cells lining vascular spaces surrounded by extensive fibroblastic proliferation and bone formation(1). Other authors mentioned injury as a pathogenic factor for ABC (4), but in our case no traumatic antecedent was known.

Table 1 Locations reported in the literature of aneurysmal bone cysts with fibrous displasia

Author	Year	REF	Location
Donaldson	1962	8	jaw
Biesker	1970	1	n. i.
Barukzewski	1971	4	rib (2 Th), jaw
Clough	1975	6	tibia
Ruiter	1977	17	n. i.
El Deeb	1980	9	jaw
Branch	1986	3	frontoparietal periorbital region
Mintz	1987	14	humerus
Martinez	1988	13	rib (5 Th)
Somm	1991	18	paranasal sinuses
Revel	1992	16	jaw

n. i.: not identified

The development of an ABC in a FD is a possibility to be considered in cases of growth of a preexisting FD to avoid it being wrongly diagnosed as a malignancy (7).

The potential for recurrence of FD is low; it is related to the age of the patient, the size of the lesion, the presence of mitoses, or the incompleteness of the resection (3).

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F. Tresserra

Department of Pathology
Hospital Mutua de Terrassa
c/Garcia Humet s/n.
E-08221 Terrassa (Barcelona)
Spain